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## Fibrous Dysplasia of Both Alae of the Sacrum

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and M. Martínez-Grande, MD

### ABSTRACT

**A case of monostotic fibrous dysplasia involving both alae of the sacrum is reported. Only 2 cases of monostotic sacral involvement were published previously. The lesion was detected in a 42-year-old man suffering from lumbosacral pain after minimal trauma. Radiographic studies revealed cystic images on both alae of the sacrum, with internal condensations and some liquid contents. A trephine biopsy did not produce enough diagnostic data, and a posterior surgical approach was elected. Curettage and refilling with allograft were performed. The pathologic anatomy study diagnosed a fibrous dysplasia. Two years after surgery, the patient was asymptomatic.**

Fibrous dysplasia is a tumorous condition primarily located in the long bones and has been described in the spine in fewer than 25 cases,<sup>1-12</sup> most of which were polyostotic forms. Its location in the sacrum is rarer still; this has been described in only 5 cases,<sup>10,13,14</sup> and 2 of them were monostotic.<sup>10,14</sup> Monostotic fibrous dysplasia with involvement of both alae of the sacrum, such as in the case presented here, must therefore be regarded as exceptional.

### CLINICAL CASE

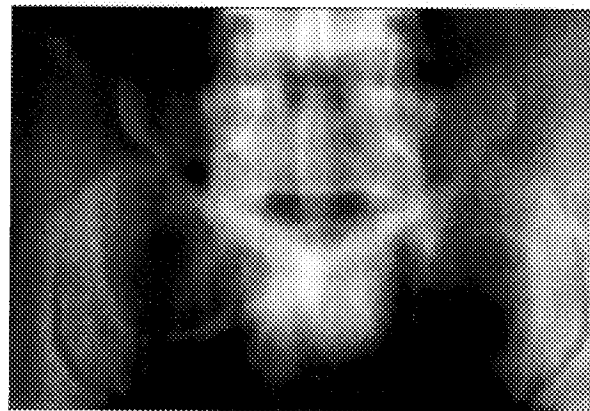
We present the case of a 42-year-old man suffering from lumbosacral pain with no sciatic involvement. Pain developed as a result of banal trauma approximately 2 years before the patient came to our hospital.

Neurological examination results were normal. Blood tests revealed no significant anomalies. Simple roentgenograms and tomography revealed lytic images on both alae of the sacrum (Figure 1). Computed tomography (CT) scans and magnetic resonance imaging (Figures 2 and 3) permitted adequate evaluation of the extent of the injuries, revealing liquid contents and internal bone trabeculation on the left side. Isotopic technetium studies

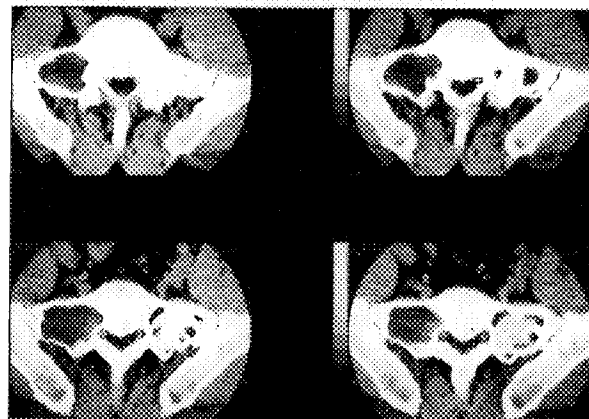
showed a minimal increased capture at the level of both alae of the sacrum.

A trephine biopsy under CT scan control did not supply enough data for diagnosis. A posterior surgical approach revealed cystic lesions with small amounts of internal serohematic liquid and solid contents in the left side. The contents were curettaged, and the cavities were filled with allograft from 4 frozen femoral heads.

The pathologic anatomy was similar on both sides and consisted of fibrous injuries with curved spicules and bone metaplasia. The spicules were in the form of osteoid metaplasia with no laminae or osteoblastic lining and were consistent with fibrous dysplasia.



**Figure 1.** Tomography cross-section showing cystic images on both alae of the sacrum in a 42-year-old man suffering from lumbosacral pain with no sciatic involvement.



**Figure 2.** Computed tomography scan study to evaluate the extent of the cystic lesions in the patient described in Figure 1—showing a clearly higher degree of bone condensation on the left side.

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Figure 3. (A) T<sub>1</sub> and (B) T<sub>2</sub> sequences of a frontal magnetic resonance imaging cross-section in the patient described in Figure 1—showing liquid and solid contents within the cysts.

The patient has recovered well, is free of pain, and remains asymptomatic 2 years after surgery.

### DISCUSSION

Fibrous dysplasia is considered to be a hamartoma and thus a congenital condition.<sup>1</sup> When associated with white-coffee-colored skin pigmentation, skeletal maturity, and early puberty (in girls), it is known as Albright's syndrome, which accounts for fewer than 5% of cases. The condition can be monostotic or polyostotic. Polyostotic forms appear between ages 5 and 20, whereas monostotic or oligostotic forms usually remain asymptomatic until adulthood.<sup>1,10</sup> Involvement of both alae of the sacrum in our case supports the congenital origin of the tumor.

In monostotic forms, the most commonly affected bone is the maxilla, followed by the femur. Monostotic forms in the spine are rare.<sup>1-12</sup> Fibrous dysplasia seldom develops in the sacrum and is involved in about 5% of cases of Albright's syndrome.<sup>15</sup> Other than in Albright's syndrome, sacral involvement is exceptional, and we have found only 3 references to it in the literature: in papers by Ehara and coworkers, who described 2 polyostotic cases,<sup>13</sup> Firat and Stutzman, who described 2 cases (1 monostotic, 1 polyostotic),<sup>14</sup> and Schajowicz, who described 1 case.<sup>10</sup> Thus, only 2 monostotic cases were described previously.<sup>10,14</sup>

Monostotic forms are normally asymptomatic, unless compression of the neural elements, microfractures, or pathologic fractures occur.<sup>1,8</sup>

Image studies show a lytic area with occasional internal opacities when the cartilage is ossified.<sup>1</sup> Differential diagnosis in the spine and the sacrum must be determined in the lytic forms for hemangioma, giant-cell tumor, and aneurysmal bone cyst. In forms with blastic predominance, osteoblastoma and Paget's disease should be ruled out. In young

patients, the possibility of multiple myeloma or metastasis must also be discounted.<sup>13</sup>

Treatment, and even biopsy, is not generally required, because image studies are highly typical, and symptoms tend to resolve spontaneously, as Campanacci and Ruggieri found.<sup>1</sup> However, when pain persists, or when the diagnosis is doubtful, as in our case, treatment might be indicated. Given that the involvement of the sacrum, and especially of both alae, is extraordinary, we were obliged to perform an open biopsy. We also decided, based on the radiological signs of benignness, to perform a curettage and allograft refill in the same operation. Nevertheless, it was appropriate to have the pathologic study results before performing definitive treatment. Most authors prefer to use autologous graft,<sup>1,4-7,9,12-14,16,17</sup> although others use allograft<sup>18-20</sup> or hydroxyapatite.<sup>6</sup> In our case, we chose allograft because of the large extent of both cavities, which could not be filled with autologous graft from the iliac crest. Given the nature of the tumor, it was not deemed necessary to perform radical resection surgery on the sacrum.

Curettage and refill grafting normally cure the condition. Some authors have described resorption after grafting,<sup>3,16,21,22</sup> but this is more frequent in young patients<sup>1</sup> and in cases in which the lesion has been incompletely excised or inadequately immobilized.<sup>7</sup> In cases in which the tumor is particularly aggressive or involves the vertebrae, most authors favor radical excision of the lesion.<sup>4,7</sup> An additional surgical indication includes cases of neurologic decompression and spine alignment procedures.<sup>1-9,11-13,22</sup>

Sarcomatous degeneration occurs in fewer than 1% of cases, both in monostotic and polyostotic forms, and is more frequent in lesions that have undergone previous radiation treatment.<sup>1,23-26</sup>

## CONCLUSION

In summary, fibrous dysplasia should be considered in the differential diagnosis of the benign lesions located in the sacrum, even when signs of polyostotic fibrous dysplasia are not present.

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